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Pulmonary Blood Supply in Patients With Pulmonary Atresia and Ventricular Septal Defect

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The heart and lung specimens in 31 cases of pulmonary valve atresia and ventricular septal defect were studied at autopsy. Three types of natural arterial blood supply to the lungs were identified: 1) ductus arteriosus (patent or ligamentous) (12 cases); 2) major collateral arteries (20 cases); and 3) diffuse small pleural arterial plexus coexisting with either ductus arteriosus or major collateral arteries (17 cases). The ductus arteriosus and major collateral arteries did not coexist in the same lung in these cases. Confluent central pulmonary arteries were present in 22 (71%) of the 31 cases, involving 7 (58%) of the 12 cases of ductus arteriosus, 14 (70%) of the 20 cases with major collateral arteries and 1 case with an aorticopulmonary window. The pulmonary trunk (atretic or patent) was identifiable in 24 (77%) of the 31 cases. A lung or lungs that connected to a ductus (or ligamentum) had a complete and unifocal intrapulmonary arterial distribution (without arborization abnormalities). Major collateral blood supply was frequently multifocal and associated with arborization abnormalities. The size of the central pulmonary arteries was not related to the type of arterial blood source but seemed to be related to the amount of blood flow actually reaching the vessels.

This study demonstrated a complex systemic arterial system supplying the lungs in these cases. The size, sources and relation among the ductus, the pulmonary artery confluence, the large and small collateral vessels and the intrapulmonary system are far more varied than has ever been reported previously. Careful and thorough premortem studies are crucial if surgical intervention is contemplated.

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The combination of pulmonary valve atresia with ventricular septal defect and biventricular origin of the aorta (generally considered a severe form of tetralogy of Fallot) is characterized anatomically by the absence of a patent direct connection between the right ventricle and any part of the pulmonary artery tree (1,2). The blood supply to the lungs is derived entirely from the systemic arterial circulation. Various terms have been used for this condition, including pseudotruncus arteriosus, pulmonary (valve) atresia with ventricular septal defect, tetralogy of Fallot with pulmonary atresia and type IV persistent truncus arteriosus (3,4).

In pulmonary atresia with ventricular septal defect, the pulmonary blood supply arises from the ductus arteriosus or major systemic to pulmonary collateral arteries or from both (2,5–8). The collateral vessels may be relatively large arteries (3 to 20 mm in diameter), varying from one to five in number and arising most commonly from the descending thoracic aorta and less commonly from the subclavian arteries, the abdominal aorta or its branches or the left coronary artery (5,9–12). A third type of blood supply to the lungs, described by Jefferson et al. (10), comprises numerous, uncountable, small systemic collateral arteries that either follow the bronchi or spread over the pleurae; others (5,13,14) have considered these an enlarged bronchial arterial circulation.

The present study was undertaken to determine the variety and frequency of the natural sources of blood supply to the lungs in autopsy cases of tetralogy of Fallot and pulmonary valve atresia. The intrapulmonary distributions of the pulmonary arteries and the various types of systemic collateral arteries were studied. Histologic study of these vessels and the peripheral pulmonary arteries was also done.

It was hoped that this study would allow certain speculations concerning the collateral vessels, including their or-

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igin, their ability to function as true pulmonary arteries and their natural history in terms of providing a reliable blood supply to the lungs.

Methods

Study cases. The study material included the clinical data and heart-lung autopsy specimens in 31 cases of ventricular septal defect and pulmonary valve atresia seen between 1975 and 1982. The ages of the patients ranged from 16 days to 30 years (mean 9.5 years), and 14 were older than 10 years of age. Two patients had died without surgical intervention, 15 had had corrective surgery with or without preceding palliative surgery and 14 had had a palliative procedure only.

Pathologic study. Postmortem gross dissection of the arterial supply to the lungs was done, and specific attention was given to identification of the pulmonary trunk, the ductus arteriosus and the central pulmonary arteries or their branches. Central pulmonary arteries were defined as extrapulmonary vessels that could be identified as either the right or the left pulmonary artery. All extrapulmonary portions of the grossly visible systemic collateral arteries that entered the lungs, including the bronchial arteries, were identified. Further dissection was then carried out into the hilus and the pulmonary parenchyma to identify all intrapulmonary arteries to the level of segmental or subsegmental bronchi. A search was made for anastomoses between the systemic collateral arteries and the pulmonary arteries, both outside and inside the lungs. An example of the dissections is illustrated in Figure 1.

The central and intrapulmonary arteries and the systemic collateral arteries were evaluated in microscopic sections, which were stained with hematoxylin-eosin and with elastic-van Gieson (to allow differentiation of elastic and muscular arteries). The presence of arterial lesions, such as intimal fibrosis or luminal thrombosis, was also recorded.

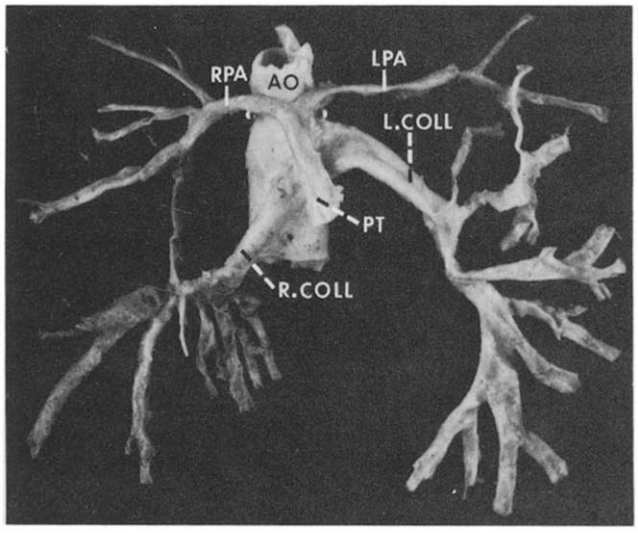

Figure 1. Dissected specimen showing two major left and right collateral arteries (L.COLL and R.COLL) originating in a common orifice on the ventral aspect of the upper descending thoracic aorta (AO). The right collateral artery anastomosed with the right lower lobe branch of the right pulmonary artery (RPA), while the left collateral artery had no connection with the central pulmonary arteries but was in continuity with the lower left intrapulmonary branches of the distal left pulmonary artery (LPA) system. PT = main pulmonary trunk.

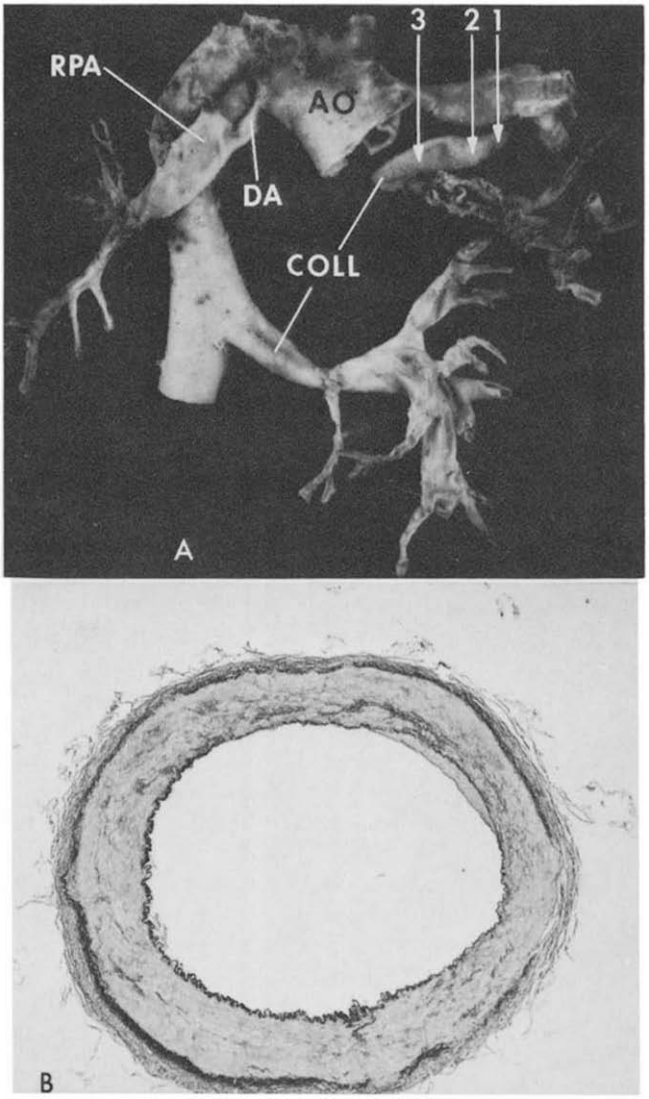

Figure 2. A, Dissection showing large redundant collateral artery (COLL), mimicking a left ductus arteriosus, which supplied a portion of the left upper lobe only. The distal right pulmonary artery (RPA) arose from a right ligamentum arteriosus (DA). Numbers 1, 2 and 3 refer to sites of sections taken for histologic study. **B,** Histologic structure of vessel sectioned at point 1 of **A.** The vessel is a thick-walled muscular systemic artery differing from the usual appearance of a ductus arteriosus (elastic-van Gieson; ×5, reduced by 20%).

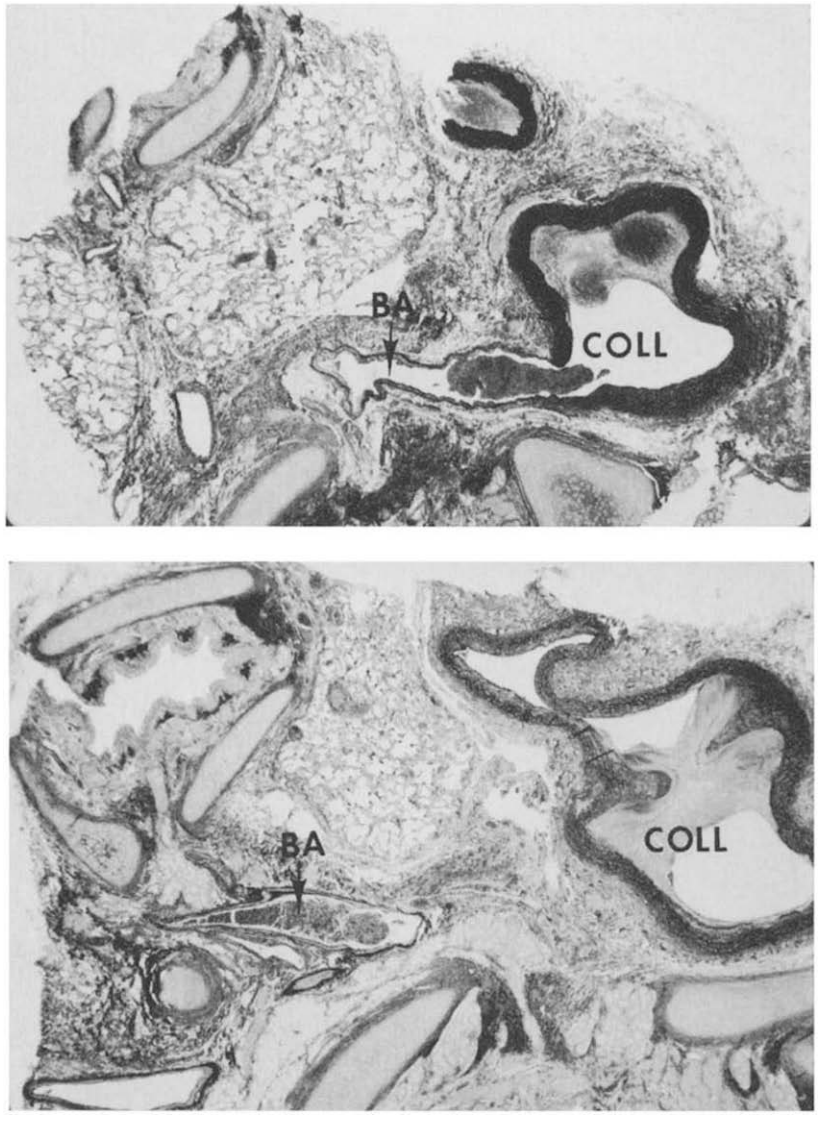

Figure 3. Small nutritive branches of a muscular bronchial artery (BA) originating in an elastic hilar branch of a major collateral vessel (COLL) in serial sections (elastic-van Gieson; × 5, reduced by 13%).

Results

Three types of systemic arterial blood supply occurred in various combinations and supplied the lungs in the 31 cases. These types were ductus (or ligamentum) arteriosus (12 cases), major systemic to pulmonary collateral arteries (20 cases) and diffuse small arteries forming a vascular plexus on the pleural surface (17 cases). The lungs were supplied by a single type of systemic source in 14 cases and by more than one type in the remaining 17 cases. In one case, there was an aorticopulmonary window with an atretic pulmonary trunk, confluent right and left pulmonary arteries and normal peripheral pulmonary artery distribution.

Ductus arteriosus. A ductus arteriosus was present in 12 cases and was ligamentous in 6 and patent but stenotic in 6; bilateral ductus were not seen in any case. The central pulmonary arteries were confluent in seven cases and congenitally nonconfluent in two, with the right lung supplied by a ductus and the left supplied by major systemic collateral arteries. In three cases, nonconfluent pulmonary arteries presumably were acquired secondary to a previous Waterston anastomosis. The ductus (or ligamentum), when present, joined the homolateral pulmonary artery and supplied either the homolateral lung alone (in the setting of nonconfluent pulmonary arteries) or both lungs (when confluent pulmonary arteries were present).

In 22 of the 24 lungs among the 12 cases with a ductus arteriosus, the pulmonary arterial distribution was normal and no major collateral vessels were present. The remaining two lungs were supplied by major systemic arterial collateral vessels. Histologically, each ductus was a thick-walled structure with a typical dysplastic appearance.

In four cases, major collateral vessels were incorrectly considered ductus by gross examination (Fig. 2A). Collateral arteries arose from the undersurface of the left aortic arch immediately distal to the origin of the left subclavian artery in two cases and from the base of the left subclavian artery in the presence of a right aortic arch in two cases.

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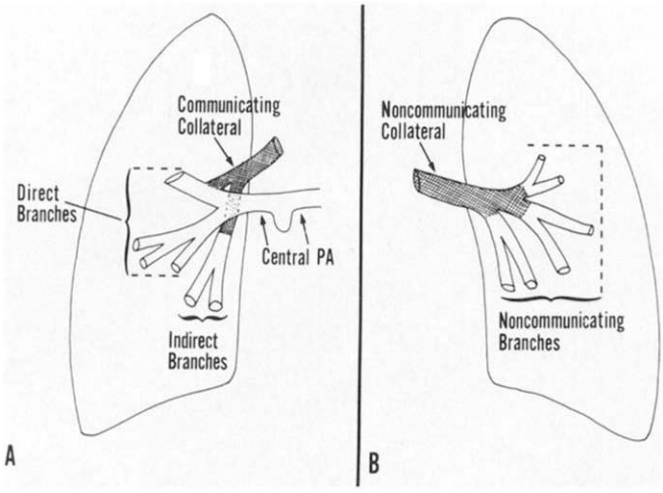

Figure 4. Schematic drawing of types of major collateral arteries seen in this study. **A,** Collateral artery that communicates directly with branches of pulmonary arterial tree, which originated in the central pulmonary artery (PA). Direct and indirect branches result from this relation. **B,** Collateral artery that communicates with intrapulmonary arterial tree without any direct communication to recognizable major true pulmonary arteries. **A** shows a communicating collateral and **B** shows a noncommmunicating collateral artery. Portions of the lung supplied by a noncommunicating collateral artery were considered arborized rather than sequestrated.

However, histologic studies suggested that these vessels were systemic collateral arteries (Fig. 2B). Moreover, the vessels had additional features that were not compatible with a ductus, such as pronounced tortuosity or bifurcation before the vessels joined the pulmonary arteries. Unlike the true ductus, which invariably connected with the pulmonary artery that supplied all segments of the lung, these ductus-like major collateral arteries always supplied only a portion of the lung.

Major collateral arteries. Eighty major collateral arteries in the 20 cases were identified and studied. In each of the 20, two to six major collateral arteries were seen sending branches to the lung parenchyma, although some also sent tiny nutrient branches to the bronchial walls and hilar lymph nodes (Fig. 3). The major collateral arteries originated from the mid-descending thoracic aorta near the level of the carina in 50 (63%), the undersurface of the aortic arch in 9 (11%), the subclavian artery in 5, the distal descending thoracic aorta in 4, the left coronary artery in 2, the abdominal aorta in 1 and unknown sites in 9. The size of the major collateral vessels ranged from 1.5 to 20 mm in outer diameter at their origins (or at the postoperative stumps in collaterals of unknown origin).

Confluent central pulmonary arteries were present in 14 (70%) of the 20 cases. In the six cases with nonconfluent central pulmonary arteries, the pulmonary trunk continued as the left pulmonary artery in one case and as the right pulmonary artery in another case. The pulmonary trunk was absent or atretic in the remaining four cases.

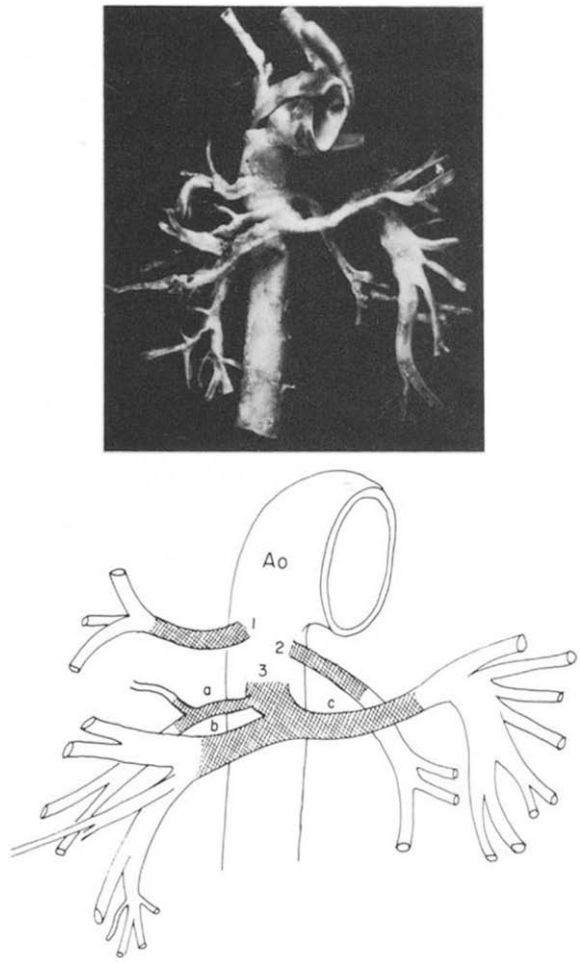

Figure 5. Dissection and schematic drawing of example of multifocal collateral blood supply in absence of confluent central pulmonary arteries. Collateral arteries 1, 2 and 3a, b and c originating in the aorta (Ao) supplied five different portions of the lung. The blood supply of the entire lung represents an arborization abnormality.

Thirty-two (40%) of the 80 major collateral arteries joined the central pulmonary arteries or their lobar or segmental branches (communicating collateral arteries) and thereby afforded a dual pathway (blood supply) to the peripheral pulmonary arteries (Fig. 4). In many instances, the area of anastomosis between the collateral artery and the pulmonary artery branch was characterized by discrete stenosis or tubular narrowing of mild to moderate degree. The remaining 48 major collateral arteries (60%) had no identifiable anastomoses with the central pulmonary arteries or their branches within the lungs; these were designated noncommunicating collateral arteries (Fig. 4). After entering the hilus, they coursed toward the nearest lobar or segmental airways and divided with the airways in a fashion similar to lobar or segmental pulmonary arteries, and they were supplied from part of a segment to as many as nine segments within a lung.

In the absence of central pulmonary arteries or ductal origin of the pulmonary arteries, the intrapulmonary arterial

branching pattern was abnormal (arborization abnormality) (Fig. 5); in other cases, the arborization abnormalities involved areas ranging from less than one segment to nearly eight segments in each lung, depending on the distribution of the noncommunicating collateral arteries (Fig. 6). In three cases (each with confluent central pulmonary arteries), multiple anastomoses between several major collateral and segmental branches of the central pulmonary arteries formed an interconnecting peripheral pulmonary arterial tree with full distribution to both lungs; however, the hilar branching pattern was aberrant (Fig. 7A). Loop anastomoses between the collateral arteries and the true pulmonary arteries also were observed (Fig. 7B).

In most instances, a bronchopulmonary segment was supplied exclusively by branches of either a central pulmonary artery or a noncommunicating collateral artery. However, in some instances, a single bronchopulmonary segment was supplied by both a central pulmonary artery and a noncommunicating collateral artery, although each apparently perfused different portions of the segment and had no grossly identifiable intrasegmental anastomoses (Fig. 8).

Arterial plexuses. In 17 cases, small arterial plexuses made up of small vessels were identified on the pleural surfaces. These plexuses arose from the base of the subclavian, internal mammary, upper intercostal and major collateral arteries, the descending thoracic aorta and, rarely, the left coronary artery (one case) and the phrenic artery (one case). Four of the 17 cases involved patients less than 1 year of age (1 being a 6 month old infant).

Anatomic interrelations. An inverse relation was observed between the presence of ductus arteriosus and major collateral arteries. Among the 20 cases with major collateral arteries, a ductus or its remnant was absent in 18 cases (90%). Moreover, among 12 cases with a ductus arteriosus, major collateral arteries were absent in 10 (83%); in 2, there was a ductal origin of the right lung, major collateral arteries to the left lung and nonconfluent central pulmonary arteries. Therefore, even when the ductus arteriosus and major collateral arteries coexisted in the same case, they supplied different lungs and were not present in the same lung.

The presence of the pulmonary trunk, atretic or patent, was usually associated with confluent and patent central pulmonary arteries. In 21 (88%) of the 24 cases with an identifiable pulmonary trunk, confluent central pulmonary arteries were present, and in 6 (86%) of the 7 cases without a pulmonary trunk, there were no confluent central pulmonary arteries. Accordingly, in 20 (95%) of the 22 cases with confluent and patent central pulmonary arteries, a pulmonary trunk was present.

Histopathologic findings. Sections of the major collateral arteries revealed segments of severe luminal obstruction ($>$ 75% obstruction of luminal cross-sectional area) in 8 (14%) of the 59 arteries studied. Moreover, the outer diameters of the severely obstructed segments were usually hypoplastic and smaller than those of the adjacent nonobstructed segments and, thereby, resembled an hourglass.

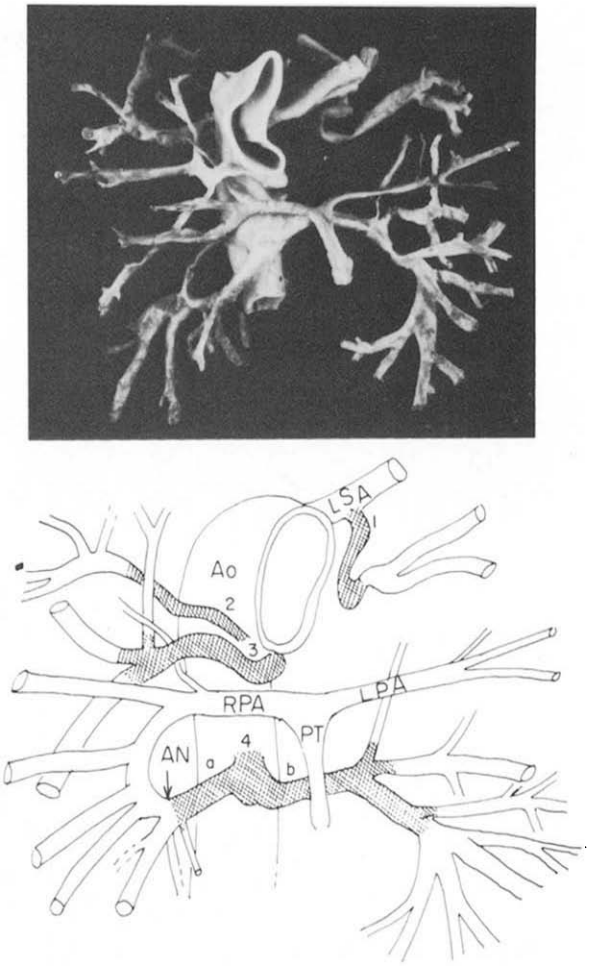

Figure 6. Dissection and schematic drawing of another example of multifocal blood supply to lungs in the presence of confluent central pulmonary arteries: the left (LPA), pulmonary trunk (PT) and the right (RPA). Noncommunicating collateral arteries 1, 2, 3 and 4b, supplying four isolated areas of lung. The rest of the lung was connected to the confluence, which obtained blood supply from collateral artery 4a. AN = systemic pulmonary arterial anastomosis as determined by histologic study; LSA = left subclavian artery. The **shaded vessels** had the histologic appearance of systemic arteries.

Histologic study of the peripheral pulmonary vascular bed revealed that, proximal to the terminal bronchioles, the transitional and larger muscular arteries often were normal-sized or only slightly hypoplastic, compared with the accompanying airways. However, the intraacinar arteries (those accompanying respiratory bronchioles or more distal airways) generally seemed to be hypoplastic, regardless of the blood source or the presence or absence of a proximal stenosis. In most cases, muscularized arteries were observed at the level of respiratory bronchioles or alveolar ducts, or both. The relative thickness of the media of the muscular pulmonary arteries varied greatly and ranged from hypertrophic to atrophic.

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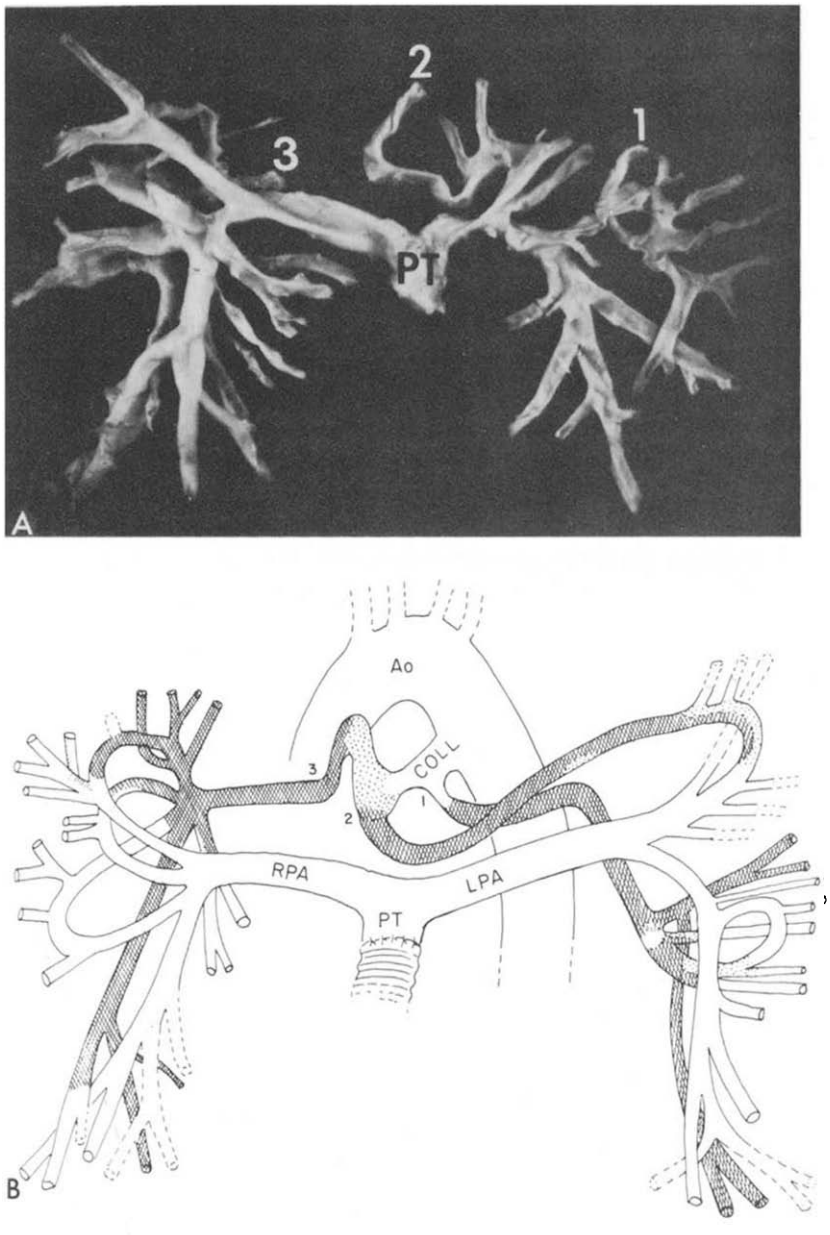

Figure 7. A, Dissection showing segmental and subsegmental branches spread out to show systemic collateral-pulmonary artery anastomosis (COLL). The branching pattern of central pulmonary arteries was abnormal at the hilar region. Numerous loop anastomoses were seen among collateral arteries 1, 2 and 3 and the branches of the central pulmonary arteries. **B,** Schematic drawing of dissection shown in **A.** The **shaded vessels** are those that appeared to be systemic arteries by histologic study. Ao = aorta; LPA = left pulmonary artery; PT = pulmonary trunk; RPA = right pulmonary artery.

Discussion

Pathologic findings. This study demonstrated the various types of systemic arterial blood supply to the lungs of patients with ventricular septal defect and pulmonary atresia that have been reported previously (2,7,12,15), but these reports did not detail the complexity of that blood supply and its relation to the true pulmonary arteries and the intrapulmonary arterial tree.

Relation of a functioning ductus to pulmonary vascular distribution. In the present study, a ductus and a major collateral artery did not coexist in the same lung. A similar finding has been described by others (2,7,15). It is also noteworthy that a functioning ductus was associated with normal distribution of the intrapulmonary vascular tree. Only very rarely has a ductal origin of a pulmonary artery been found in a lung that also received a major collateral artery (5,16). However, we observed several ductus-like collateral arteries that originated near the anticipated site of the true ductus arteriosus but differed from it histologically and grossly, having tortuosity and branching.

Because the ductus arteriosus anastomoses with proximal central pulmonary arteries outside the lung and provides adequate anterograde flow during fetal life, the central pulmonary artery may be normal sized at birth and become hypoplastic postnatally, after ductal closure or stenosis. In these lungs, a premortem diagnosis of absent pulmonary artery may be made. However, patent pulmonary arteries at the hilar area, supplied by alternative sources, can always be identified at autopsy (17). Our findings support this concept.

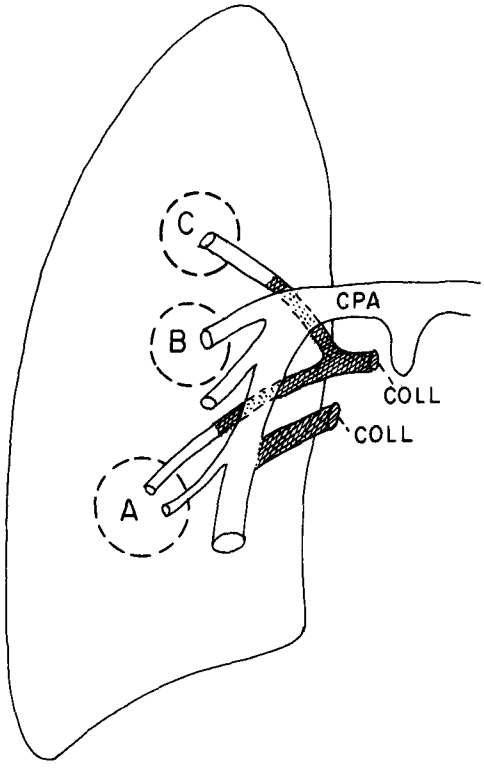

Figure 8. Schematic drawing of the parallel supply and sole supply within a bronchopulmonary segment defined at autopsy. Parallel blood supply is shown in **A.** Two types of sole supply are shown in **B** and **C.** CPA = central pulmonary artery; COLL = the collateral arteries.

Relation of central pulmonary and major collateral arteries. Although some investigators (10,15,18) have observed that the sizes of the central pulmonary and systemic collateral arteries are inversely related, others (2,11,14,19) have reported normal-sized central pulmonary arteries in lungs supplied by large collateral arteries, and this was also observed in our study. Our study and others (2,15,18–20) suggest that once the central pulmonary arteries are formed during fetal life, their size is most likely determined by the amount of blood flowing through them rather than by their congenital source of blood supply.

Multiple major collateral arteries that anastomosed with central pulmonary artery branches at segmental or subsegmental artery levels in multiple sites were associated with very hypoplastic central pulmonary arteries. In contrast, central pulmonary arteries supplied by major collateral arteries with anastomoses at the lobar artery level or outside the lung (for example, coronary-pulmonary artery fistula) showed less hypoplasia or were even normal sized.

In contrast to the size of the central pulmonary arteries, the distribution of the intrapulmonary vascular tree was determined by the types of congenital blood supply. Because a ductus arteriosus usually does not coexist with major collateral arteries in the same lung, ductal supply was associated with normal peripheral distribution within the lungs. Major collateral arteries were frequently associated with multifocal blood supply to the lung and with arborization abnormalities.

The coexistence of small diffuse and major collateral arteries was observed in our study. Some stenotic major collateral arteries actually gave rise to these small pleural arterial plexuses. We are not aware of similar findings in other studies, with the exception of one case in the series reported by Allanby et al. (5). We agree with others (10,14,19,21) that this type of collateral pattern is probably acquired postnatally in response to regional flow reduction, such as in ductal stenosis, stenotic surgical shunt or stenotic major collateral vessels, rather than in response to systemic hypoxia. This view is supported by the observation of Rabinovitch et al. (12) in patients with tetralogy of Fallot.

Clinical implications. Previous studies (5,6,20,22) had shown that the ductus arteriosus is a precarious sole source of pulmonary blood supply. In our series, 60% of the cases with a ductal source but no major collateral arteries had required a surgical shunt in early infancy, and severe hypoxia requiring surgical intervention developed in the remaining patients before early childhood. Major collateral arteries are a more stable source of pulmonary blood supply, probably because of their multiple numbers. Although pulmonary blood flow may be excessive in some patients (3,19,20,23), a high incidence of major collateral vessel stenosis has been observed, and significant obstruction may progressively develop, starting even in infancy (12,22,24). Despite sporadic reports of unusual longevity of these patients, major collateral arteries probably cannot provide adequate flow for a normal life span in most instances (22).

The status of the peripheral pulmonary vascular bed is difficult to assess in these patients (12). Different portions of the lung in this anomaly may show different structural changes, according to their source of blood supply. Both hyperperfusion and hypoperfusion, regardless of the type of blood supply, cause structural abnormalities that may affect pulmonary resistance (12,25,26). The problem is further complicated because the diameters and total numbers of the intraacinar arteries may be reduced in this anomaly (12,27).

Surgical implications. Because blood flow is the key factor for pulmonary artery growth, early surgical intervention to ensure adequate blood supply to the central as well as peripheral pulmonary arteries is often necessary. An early palliative operation is often chosen for patients with hypoplastic central pulmonary arteries (14,19,20), because immediate dilation of the hypoplastic central pulmonary arteries often follows a first stage palliative operation or central shunt, and progressive enlargement of the arteries may continue postoperatively (19,28–30). The size of the central pulmonary arteries can sometimes be acutely distended at surgery by increasing the perfusing pressure and flow.

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Chronically, the growth of the central pulmonary arteries is probably stimulated by the flow that actually reaches the central pulmonary arteries, whether anterograde (through a ductus arteriosus or, rarely, a major collateral artery or through a surgically created right ventricular outflow or central shunt) or retrograde (through a major collateral artery or a surgical shunt).

Surgical experience so far seems to suggest that postoperative pulmonary artery resistance is determined primarily by the size of the left and right pulmonary arteries (11), although arborization abnormalities and stenoses of the hilar or segmental arteries may have a definite but as yet unmeasurable role.

Functional role of the collateral arteries. Major collateral arteries, whether they anastomose with central pulmonary arteries or their branches or are the sole supply to an intrapulmonary artery system, indirectly participate in alveolar gas exchange. However, their nutritive function as normal bronchial arteries has been questioned (10,12). Our study demonstrated that small nutritive branches arise from collateral arteries. To avoid possible pulmonary infarction, this nutritive function should be considered before a noncommunicating collateral artery is ligated at its origin.

Although the evidence is inconclusive on whether these collateral vessels are true bronchial arteries, we believe that they probably are persistent intersegmental arteries which are less differentiated than the conventional bronchial arterial tree and that they are likely to grow to a large size.

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